CASE REPORT

Gross anterior segment ischaemia following vitreoretinal surgery for sickle-cell retinopathy

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SUMMARY

We report the case of a 32-year-old Afrocaribbean man with known stage 3 proliferative sickle-cell retinopathy who presented with a mixed picture of tractional and rhegmatogenous macula off detachment. He underwent left primary 25 g vitrectomy with silicone oil, delamination and endolaser photocoagulation under a general anaesthetic. He, however, presented 48 hours postoperatively with gross anterior segment ischaemia. His pain and ocular signs settled over the course of a few days following administration of supplemental oxygen, oral steroids, analgesia and intravenous hydration. Examination showed resolution of his proptosis and orbital signs as well as anterior segment inflammation. He remains under follow-up.

BACKGROUND

Nucleotide abnormalities in the β -chain of haemoglobin manifest as sickle-cell disease which leads to cellular dehydration and vaso-occlusive events throughout the body. This is stimulated by pathophysiological mechanisms that are not fully understood that include hypoxia, hypotension and inflammation.¹ There is an estimated incidence of 300 000 sickle-cell births each year, with the highest prevalences being recorded within Sub-Saharan Africa, India, the Middle East and the Mediterranean.² Emigration has led to around 100 000 individuals living with the disease within the USA and 10 000 within the UK, a figure expected to rise.^{2,3} General management and prevention of systemic sequelae of sickle-cell disease focuses on treatment with hydroxyurea, blood transfusion, vaccination, stem cell transplant and antibiotic therapy.²

Ophthalmic sickle-cell disease has a variety of manifestations, representing a spectrum of ocular ischaemia, with haemoglobin C and beta thalassaemia mutations said to be associated with the worst cases of retinopathy. Potential complications include orbital wall ischaemia, anterior segment ischaemia (ASI), sectoral iris atrophy, rubeosis iridis, macular holes and proliferative sickle retinopathy (PSR). PSR comprises peripheral artery occlusion, arteriovenous anastomoses, neovascular and fibrous proliferations (sea fans), vitreous haemorrhage and retinal detachment.³

Retinal detachments occur due to mixed mechanisms, often with both tractional and rhegmatogenous components. Historically, scleral buckling procedures have been used with high incidences of postoperative ASI.⁴ Nowadays, pars plana vitrectomy (PPV) is advocated for treatment of retinal detachment with good

outcomes reported from fairly large case series.^{5,6} Unlike scleral buckling, PPV is not thought to be associated with ASI. We report a rare case of this occurring in a patient undergoing PPV for retinal detachment secondary to sickle-cell disease.

CASE PRESENTATION

We report a case of a 32-year-old Afrocaribbean man who had been attending the medical retina clinic for over a period of 4 years. He had been noted to have stage 3 proliferative changes in areas of his right eye which prompted treatment with unilateral sectoral panretinal photocoagulation laser. His ocular condition was stable with 6/6 snellen acuity in both eyes and he remained asymptomatic. His past medical history included a previous Transient Ischaemic Attack (TIA) a few years earlier that was attributed to his sickle-cell disease and for this he took clopidogrel. He had suffered no previous sickling crises.

Two years passed and after some missed appointments, he presented to our eye casualty department complaining of reduced vision and changes in his peripheral field in the left eye. Remarkably, his left snellen acuity was 6/12 with a pinhole. He was found to have a mixed picture of tractional and rhegmatogenous macula off detachment, as shown in [figure 1](#).

After discussion with the patient it was decided to proceed with left primary 25 g vitrectomy with silicone oil, delamination and endolaser photocoagulation under a general anaesthetic. A guarded prognosis in terms of visual improvement was discussed with the patient due to the chronicity of the changes. Surgery was uneventful and the patient was discharged home safely. He was reviewed the next day by the vitreoretinal team and was making a satisfactory recovery. Some anterior segment inflammation was noted, with normal intraocular pressure (IOP) and a flat retina under silicone oil.

Forty-eight hours postoperatively, the patient re-presented with severe pain and photophobia in his left eye. On examination, there was a 2 mm L-sided proptosis, restricted eye movement, ptosis and fixed pupil. Slit lamp examination demonstrated a hazy cornea with large descemet folds and circumferential splitting of the corneal endothelial layer. The anterior chamber was showing 2+ cells. No hypopyon was seen, IOP was 14 mm Hg and a brunescant cataract was present ([figure 2](#)).

An orbital CT scan performed that day demonstrated periorbital inflammatory changes but no orbital collection. In view of these findings, IV antibiotics and high-dose oral steroids were commenced.

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Figure 1 Left fundal image in 2014 (A) and 2017 (B).

Anterior segment optical coherence tomography revealed open drainage angles with gross corneal oedema. A course of topical and oral steroid did not worsen the clinical picture. He was reviewed by our haematology colleagues for consideration of systemic management of sickling crisis. A blood transfusion was considered unlikely to improve the situation.

DIFFERENTIAL DIAGNOSIS

Endophthalmitis: this was considered but no hypopyon was present and the clinical picture improved with topical and oral steroids.

Orbital infection: no concurrent sinus disease was noted and no collection seen on CT scanning.

Severe postoperative inflammation/toxic anterior segment: no hypopyon was seen and clinical picture was out of proportion compared with what is usually seen in this condition. Orbital signs are rare with this condition.

TREATMENT

The patient was admitted with involvement of the haematology team for systemic management. He was given supplemental oxygen, analgesia and intravenous hydration. His pain subsided and his ocular signs settled over the course of a few days. Examination showed resolution of his proptosis and orbital signs as well as anterior segment inflammation. A dense cataract remained with silicone oil in the posterior chamber.

OUTCOME AND FOLLOW-UP

He remains under follow-up as is awaiting cataract surgery and silicone oil removal.

DISCUSSION

Management of sickle-cell retinopathy is difficult, with paucity of current evidence available to guide clinicians when treating such patients. A Cochrane review of only two studies demonstrated that the use of laser photocoagulation had no significant benefit when compared with conservative management in terms of disease

regression or progression.⁷ Anti-vascular endothelial growth factor is an interesting option, although only a few case reports show it being used successfully.^{8–10} PPV appears to be the treatment of choice for retinal detachments, with a UK cohort study demonstrating good visual outcomes in seven patients undergoing retinal detachment surgery.⁶ A similar study from the USA reports eight eyes undergoing PPV, with 50% requiring secondary surgery for redetachments.¹¹ The largest cohort of such patients is a recent study from the UK reporting the outcomes of 38 eyes undergoing 20 or 23 g PPV for combined tractional and rhegmatogenous detachments. No statistically significant difference was found between preoperative and postoperative visual acuity in these patients, with a redetachment rate of around 20%.⁵ All of the aforementioned studies clearly state that no episodes of ASI were encountered following surgery, with no specific systemic considerations made in the perioperative period with regard to reducing sickling crises.

The shift towards vitrectomy for managing sickle-cell detachments was as a result of high incidences of ASI following scleral buckling.^{4,12} The first report of surgery for proliferative sickle retinopathy was published in 1971 by Ryan and Goldberg.⁴ They reasoned that the encircling procedure and surgically-induced inflammation interfered with the blood flow in the ciliary vessels sufficiently to precipitate localised sickling and ischaemia. As a result, it was recommended that preoperative management included exchange transfusion, local anaesthesia with retrobulbar anaesthetics without sympathomimetics, supplemental oxygen, avoidance of treatment or compression of long posterior ciliary vessels, careful intraocular pressure control, avoidance of excessive cryopexy and avoidance of extraocular muscle removal intraoperatively.⁴ With the increasing use of vitrectomy, the vogue shifted away from this perioperative management, with modern approaches mirrored by the previously discussed publications.^{5,6,11} Some authors even recommend actively against using transfusions.¹² A recent Cochrane review was unable to make recommendations with regard to preoperative transfusions for any kind of surgery in sickle-cell patients. The two papers used for this review showed no statistically significant difference between patients receiving a transfusion or no transfusion in terms of vaso-occlusive crises, infections or perioperative complications.¹³

Review of the literature suggests that postoperative ASI is not a major concern with a PPV approach. There is only one case report of such a complication in a patient similar to ours.¹ There is no agreed consensus as to the optimal perioperative management of such patients, with no strong evidence to suggest that the aforementioned preoperative work up is effective at reducing sickling crises. However, although this is a rare complication, it is something that the ophthalmic community should be made aware of. This is the first case to be reported from a UK centre. Increasing the awareness of such outcomes will help guide the consenting process and aid decision-making with regard to when to intervene surgically for such cases.

Figure 2 Image of anterior segment and proptosis at presentation 48 hours post operatively.

Learning points

- ▶ Anterior segment ischaemia (ASI) is more often associated with scleral buckling techniques in this group of patients.
- ▶ Although rare, ASI can occur in patients undergoing pars plana vitrectomy.
- ▶ There is no clear consensus as to how these crises can be avoided during ophthalmic surgery.
- ▶ Optimal management of a sickling crisis involves supplemental oxygen and intravenous resuscitation support. Some cases may require transfusion exchange.

Contributors All four authors, AW, BG, CC and FDS, have met or reviewed the patient in a professional setting related to his clinical care and were involved in the conception and design of this report. All authors contributed significantly to the writing of this case report and gave final approval of the version published. All agree to be accountable for the article and to ensure that all questions regarding accuracy or integrity of the article are investigated and resolved as stipulated by the ICMJE recommendations 2013. Furthermore, each author is able to identify which co-authors are responsible for specific other parts of the work however, also have confidence in the integrity of the contributions of their co-authors. There are no other individuals who fulfil the above criteria.

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